CLINICAL RESEARCH STUDY

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Incidence and Prognosis of Ventricular Arrhythmias in Patients with Congenital Left Ventricular Aneurysms or Diverticula

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ABSTRACT

BACKGROUND: Patients with congenital left ventricular aneurysms and diverticula may present with arrhythmia. The incidence of ventricular arrhythmias and the clinical outcome of these patients have not been reported to date.

METHODS: Among 250 consecutive patients with congenital left ventricular aneurysms and diverticula detected by echocardiography, the clinical outcome of patients who presented with ventricular arrhythmias or associated symptoms was investigated.

RESULTS: Of 250 patients with congenital left ventricular aneurysms and diverticula, 30 had ventricular arrhythmias or syncope at initial presentation. During a follow-up of 85 months, spontaneous ventricular tachycardia occurred in 17 of these patients (57%). Ventricular tachycardia was sustained in 13, with a monomorphic pattern in 9 patients. In 82% (11 patients), ventricular tachycardia was inducible during electrophysiologic testing. In 7 patients a sustained monomorphic ventricular tachycardia with a right bundle branch block pattern similar to the clinical tachycardia was induced. Twenty patients were treated with antiarrhythmic agents. Eleven patients received an implantable cardioverter defibrillator. Appropriate device discharges were observed in 73% during a follow-up of 61 months. One patient underwent surgical resection of a congenital left ventricular aneurysm. Three patients underwent successful catheter ablation for incessant ventricular tachycardia. Of these, 2 were free of any clinically relevant arrhythmia during follow-up. Three patients died (10, 41, and 89 months after initial presentation). In 2 of them, the cause of death was attributed to ventricular arrhythmia.

CONCLUSION: The clinical outcome of patients with congenital left ventricular aneurysms and diverticula and arrhythmia is variable. Clinical ventricular tachycardia in these patients is often monomorphic and usually inducible during electrophysiologic study, indicating a role for this test in risk stratification. Appropriate discharges are frequent in implantable cardioverter defibrillator recipients with congenital left ventricular aneurysms and diverticula.

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Congenital left ventricular aneurysms and diverticula are rare disorders that seem to occur owing to disrupted embryogenesis beginning as early as the fourth week of embryonic life.[1] The diagnosis is established using cardiac imaging studies, such as transthoracic echocardiography or left ventricular angiography, after exclusion of coronary artery disease, cardiomyopathies, and other inflammatory or traumatic causes.[2-7] Most patients with congenital left ventricular aneurysms and diverticula are clinically asymptomatic. However, patients may present with cardiac arrhythmias, embolic complications, or heart failure.[8]

There are only few observational case reports and small series on congenital left ventricular aneurysms and diverticula in the medical literature.[9-14] The association of this abnormality with ventricular tachyarrhythmia was reported in 1971 by Maloy et al.[9] However, very little is known on the clinical outcome of these patients to date. In this study, we aimed to investigate the long-term outcome of patients with congenital left ventricular aneurysms and diverticula who had ventricular arrhythmias or presyncope/syncope at initial presentation.

CLINICAL SIGNIFICANCE

- Ventricular tachycardia in patients with congenital left ventricular aneurysm or diverticula is often monomorphic and has a right bundle branch block morphology.
- In patients with congenital left ventricular aneurysm or diverticula, the clinical ventricular tachycardia is usually inducible during electrophysiologic study, indicating a role for this test in risk stratification.
- In some patients the implantation of an implantable cardioverter defibrillator is required to prevent life-threatening ventricular tachyarrhythmias.

METHODS

We retrospectively screened hospital charts dated 1990 and after for patients diagnosed with congenital left ventricular aneurysms and diverticula in the Clinic for Cardiology, University Hospital, Zürich, Switzerland. The diagnosis of congenital left ventricular aneurysms and diverticula was made using transthoracic echocardiography after exclusion of coronary artery disease, local cardiac inflammatory process, or traumatic causes, as well as cardiomyopathies in all patients. Patients with diverticula or aneurysms in cardiac chambers other than the left ventricle were also excluded from the analysis.

All patient records were screened for demographic and clinical data, 12-lead echocardiogram (ECG) findings, transthoracic ECGs, 24-hour Holter recordings, and electrophysiologic studies, if available. Data available from hospital charts and those obtained from referring physicians on the clinical management of these patients, including pharmacologic treatment, catheter ablation, and implantation of an implantable cardioverter defibrillator, were collected. The clinical outcome of these patients was assessed until the date of last follow-up.

On transthoracic echocardiography, a left ventricular *aneurysm* was diagnosed in the presence of a protrusion from the left ventricular cavity, typically with a wide base, not necessarily contracting in synchrony to the left ventricle (depending on the amount of myocardial involvement). In contrast, a left ventricular *diverticulum* was characterized by a finger-like protrusion with a narrow base, which contracts in synchrony with the ventricular chamber.

All patients in the study cohort presented clinically with arrhythmic manifestations at baseline. This included ECG documentation of ventricular arrhythmias or ectopic beats, or clinical symptoms suggesting the presence of an underlying arrhythmia, such as palpitations or presyncope and/or syncope.

RESULTS

A total of 250 patients had the diagnosis of congenital left ventricular aneurysms and diverticula in our institution. In this cohort of patients, 30 had ventricular arrhythmias or presyncope and/or syncope at initial presentation. Characteristics of these patients are provided in **Table 1**. The study cohort had a mean (±SD) age of 46.6 ± 19.4 years. Twenty-one patients (70%) were male. Two patients had a family history of sudden death in the absence of a known cardiac disorder (patients 8 and 20).

On transthoracic ECG, the mean left ventricular ejection fraction was 57% ± 8.7%. The congenital left ventricular aneurysms and diverticula had a posterobasal localization in 12, apical in 9, anteroseptal in 4, and anterolateral in 5 patients (**Figures 1** and **2**). Six patients presented with more than 1 left ventricular aneurysm and diverticulum.

Arrhythmic Manifestations at Presentation

One patient had survived sudden cardiac death with documented ventricular fibrillation at initial presentation (patient 5). Seven patients (23%) presented with a ventricular tachycardia at baseline. Six patients (20%) had symptomatic ventricular ectopic beats. The remaining patients presented with presyncope and/or syncope.

Arrhythmias During Follow-Up

Multiple 12-lead ECGs and 24-hour Holter recordings revealed spontaneous ventricular tachycardia in 17 patients (57%) with congenital left ventricular aneurysms and diverticula; 13 of these had a sustained ventricular tachycardia. The ventricular tachycardia was monomorphic in 9 patients (**Figure 3**), polymorphic in 4 patients, and both monomorphic and polymorphic in 1 patient. Eight patients

Table 1 Electrophysiological Findings and Therapies in Patients with Congenital Left Ventricular Aneurysms and Diverticula Who Had Arrhythmic Manifestations at Initial Presentation

Patient No.	Age, y	Gender	Spontaneous Arrhythmias and/or Inducibility on EPS, if Available	LVA/D Location	Drug Therapy	Abl/ICD/Surgery
1	65	M	VT (on monitor)	A	ACEI, OAC	None
2	34	M	VES (LSB pattern, inf. axis)	AL	None	None
3	40	M	Syncope	AS	None	None
4	49	M	Syncope	A	BB, ASS	None
5	39	M	VF	PB	None	ICD
6	76	M	Ns VT	A	None	None
7	68	M	Monomorphic VT and/or EPS negative	PB	BB	ICD
8	34	F	Ns polymorphic VT and/or same VT on EPS	PB	BB	ICD
9	56	F	Ns polymorphic VT and/or same VT on EPS	PB	None	None
10	20	F	Syncope	AS	None	None
11	30	M	Polymorphic VT and/or Same VT on EPS	A	None	ICD
12	78	F	VES (RBB pattern, sup. axis) and/or EPS negative	AL	BB	None
13	35	M	VT (RBB pattern, inf. axis) and/or Same VT on EPS	PB	None	Abl
14	58	M	VT (RBB pattern, inf. axis)	AS	Amio, flecainide/BB	ICD
15	21	M	VT (RBB pattern, inf. axis) and/or VF induced on EPS	AL	None	ICD
16	22	M	Syncope	AL	None	None
17	72	M	None and/or Polymorphic VT on EPS	PB	Amio, ACEI	None
18	28	M	Syncope	AS	None	None
19	19	F	VT (RBB pattern, sup. axis) and/or Same VT on EPS	PB	None	None
20	48	F	Ns VT	A	BB	None
21	21	M	Syncope	A	None	None
22	63	M	VES on Holter	A	BB, ACEI	None
23	62	M	VES (LBB pattern, inf. axis)	A	None	None
24	70	M	Sustained VTs	PB	Digoxin, chinidin	ICD, surgical resection
25	62	M	Polymorphic VT and/or Same VT on EPS	PB	BB, ACEI	Abl, ICD
26	23	F	VES (LBB pattern, inf. axis)	A	None	None
27	58	M	VT (RBB pattern, sup. axis)	PB	None	None
28	49	F	Polymorphic VT	PB	BB, ACEI, Amio, Mex	ICD
29	28	M	VT (RBB pattern, sup. axis) and/or Same VT on EPS	AL	BB, Amio	Abl, ICD
30	69	F	VT (RBB pattern, sup. axis)	PB	BB, ACEI	ICD

A = apical; Abl = ablation; ACEI = angiotensin-converting enzyme inhibitor; AL = anterolateral; Amio = amiodarone; AS = anteroseptal; ASS = aspirin; BB = β-blocker; EPS = electrophysiologic study; F = female; ICD = implantable cardioverter defibrillator; inf. = inferior; LBB = left bundle branch; LVA/D = left ventricular aneurysms and diverticula; M = male; Mex = mexiletine; Ns = nonsustained; OAC = oral anticoagulation; PB = posterobasal; RBB = right bundle branch; sup. = superior; VES = ventricular extrasystole.

had ventricular tachycardia with right bundle branch block morphology, whereas 2 patients had ventricular tachycardia with left bundle branch block morphology. In the remaining patients, the exact morphology could not be determined owing to lack of a 12-lead ECG recording during the arrhythmia.

Electrophysiologic Testing

Eleven patients underwent electrophysiologic testing, of whom 9 had a prior documented spontaneous ventricular tachycardia. The other 2 patients were tested for evaluation of syncope. Electrophysiologic study results were positive in 9 patients (82%). In 7 patients with a positive study, the induced sustained monomorphic ventricular tachycardia displayed a similar morphology as during the clinical arrhythmia.

Patient Management

Twenty patients (67%) were treated with antiarrhythmic agents: β-blockers in 11, amiodarone in 5, and mexiletine, quinidine, and flecainide, each in 1 patient. In addition, 7 patients received angiotensin-converting enzyme inhibitors or angiotensin receptor blockers. Eleven patients underwent implantation of an implantable cardioverter defibrillator (single-chamber device in 10, dual-chamber device in 1 patient). Indications for implantable cardioverter defibrillator therapy were survived sudden cardiac death in 1 patient and spontaneous ventricular tachycardia in the other 10 patients. Three patients underwent successful catheter ablation for incessant ventricular tachycardia (patients 13, 25, and 29). Ventricular tachycardia with a demonstrable re-entry mechanism was rendered noninducible in all 3 patients. One patient (patient 24) underwent surgical resection of a

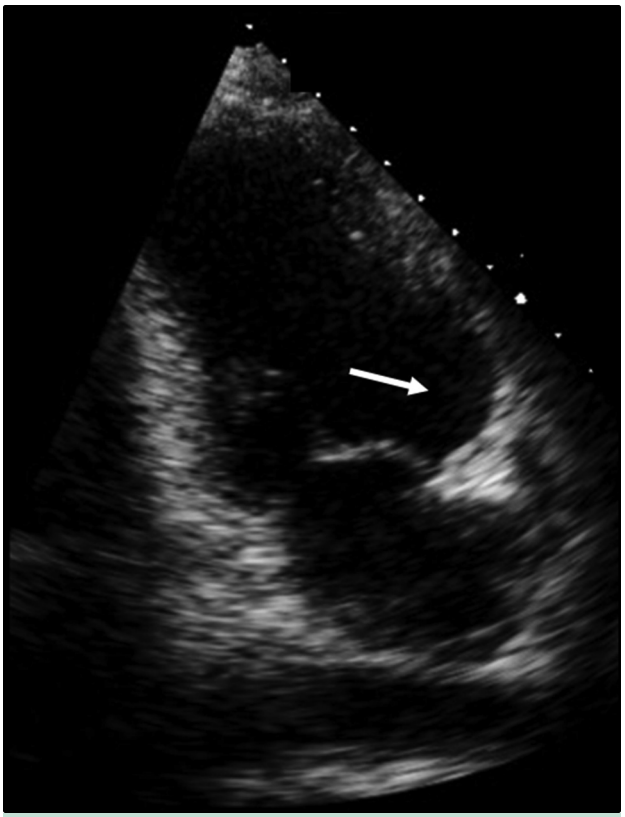

Figure 1 Transthoracic echocardiogram of a 28-year-old man (patient 29) who had syncope and repetitive monomorphic ventricular tachycardia at presentation, showing a congenital anterolateral left ventricular aneurysm (arrow) on an apical 2-chamber view.

Figure 2 Two-dimensional computerized tomogram of a 58-year-old man (patient 27) with repetitive monomorphic ventricular tachycardia (right bundle branch block morphology, superior axis) at initial presentation, showing a congenital posterolateral left ventricular diverticulum on axial view (arrow).

left ventricular aneurysm combined with a mitral valve replacement because of significant mitral regurgitation in the same year of the diagnosis.

Clinical Outcome

The study cohort had a mean follow-up of 85 ± 60 months (range 1-249 months). Three patients (10%) died at 10, 41, and 89 months after initial presentation (patients 4, 5, and 24), respectively. In 2 of these patients the cause of death was attributed to ventricular arrhythmia. Among the surviving patients, 20 patients were free of arrhythmic symptoms during follow-up. The remaining 7 patients experienced occasional arrhythmic manifestations during follow-up. Among the patients who received an implantable cardioverter defibrillator, appropriate device discharges were observed in 8 patients (73%) during a mean follow-up of 61 ± 49 months (range 1-168 months) after device implantation. The treated arrhythmia was similar to the spontaneously observed or induced arrhythmia in all of these patients. Patients who underwent catheter ablation were alive at 180, 24, and 14 months after the procedure, respectively, and 2 of them were free of any clinically relevant arrhythmia during follow-up. The patient (patient 24) who underwent surgical resection was subsequently clinically stable but died 89 months after surgery at the age of 78 years.

DISCUSSION

Congenital left ventricular aneurysms and diverticula are rare malformations with a prevalence of approximately 0.8% in patients undergoing coronary angiography.[8] The majority of these patients are free of symptoms. However, some patients may present with potentially life-threatening clinical conditions due to the presence of these disorders, such as ventricular tachyarrhythmias, congestive heart failure, and systemic embolic complications.

An association between the presence of congenital left ventricular aneurysms and diverticula and ventricular arrhythmias was observed in several reports. The initial case reported by Maloy et al was a 26-year-old woman with an apical congenital left ventricular aneurysm who had recurrent ventricular tachycardia.[9] Later, Fellow et al reported 2 patients with aborted sudden cardiac death and 1 patient who presented with syncope and documented nonsustained ventricular tachycardia.[15] Shen et al described a male patient with a sustained ventricular tachycardia caused by a posterobasal left ventricular diverticulum, which was inducible during electrophysiologic testing.[10] Likewise, Santamaria et al reported a 38-year-old woman with a posteroseptal left ventricular aneurysm and recurrent ventricular tachycardia with right bundle branch block morphology.[12] Sierra et al

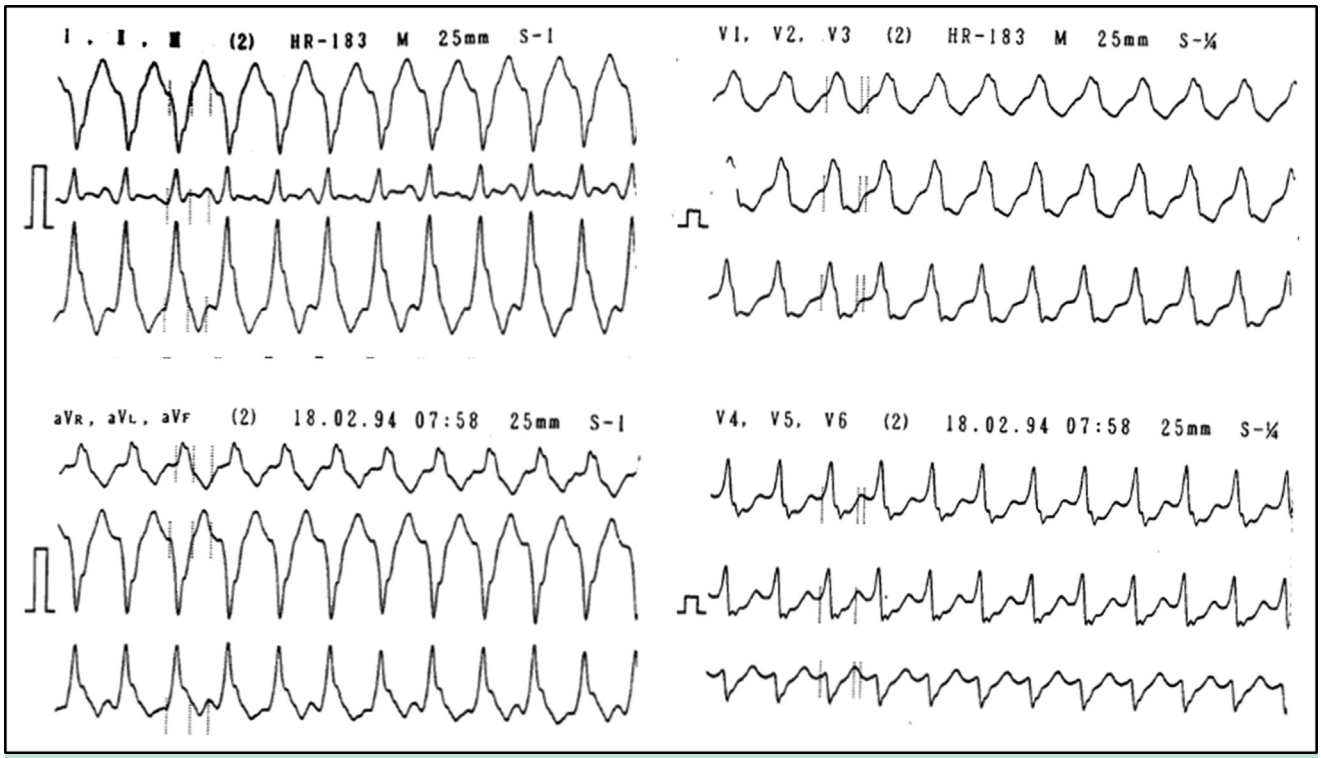

Figure 3 Twelve-lead surface electrocardiogram of a monomorphic ventricular tachycardia at a rate of 183 beats/min (right bundle branch block morphology, inferior axis, and negative deflections in leads I and aVL) of a 35-year-old man (patient 13). An echocardiogram showed a congenital posterobasal left ventricular diverticulum.

have described a 30-year-old woman with a basal left ventricular diverticulum and sustained monomorphic ventricular tachycardia.[13] Tada et al reported the electrocardiographic characteristics of ventricular tachycardia in 10 patients with congenital left ventricular aneurysms.[16] To our knowledge, our study reports clinical outcome data from the largest series of patients with congenital left ventricular aneurysms and diverticula who presented initially with arrhythmic manifestations.

We were able to demonstrate that sustained ventricular tachycardia was the most common spontaneous and inducible arrhythmia documented in patients with congenital left ventricular aneurysms and diverticula. Importantly, patients with congenital left ventricular aneurysms and diverticula and arrhythmias have a high probability of ventricular tachycardia inducibility during electrophysiologic testing, as indicated by our data. Thus, electrophysiologic testing could play a role in risk stratification in this disorder. As expected, ventricular tachycardia displayed a right bundle branch block-like morphology in most cases, suggesting a left ventricular origin of the arrhythmia.

The primary diagnostic workup in patients with congenital left ventricular aneurysms and diverticula should carefully explore the medical history for arrhythmic manifestations, such as palpitations or syncopal spells. All patients should be scrutinized for arrhythmia occurrence by ECGs and Holter recordings. If initial investigations suggest or demonstrate an underlying arrhythmia, further invasive electrophysiologic testing is warranted to assess the electrical vulnerability and risk for life-threatening ventricular tachyarrhythmia in this patient population with an underlying morphologic substrate. It is noteworthy that even though most patients with congenital left ventricular aneurysms and diverticula are clinically free of symptoms, this congenital disorder is not a benign condition, because sudden cardiac death due to ventricular arrhythmias can occur at initial presentation and during follow-up. Antiarrhythmic agents are often considered as first-line therapy. However, given the small numbers of patients, we cannot reliably comment on the efficacy of medical or device therapy in our patients. Despite the lack of guidelines for this rare disorder, implantable cardioverter defibrillator therapy should be evaluated for secondary prevention in patients who have experienced arrhythmic events as well as for primary prevention in patients with inducible sustained ventricular tachycardia or fibrillation during electrophysiologic testing. In any case, the treatment strategy must be decided on an individual basis, depending on the clinical presentations and type of arrhythmia, as well as the presence of other conventional risk factors, such as left ventricular dysfunction.

CONCLUSION

The clinical outcome of patients with congenital left ventricular aneurysms and diverticula presenting with

arrhythmic manifestations is variable. Ventricular tachycardia in patients with congenital left ventricular aneurysms and diverticula is often monomorphic and has a right bundle branch block morphology. Clinical ventricular tachycardia is usually inducible during electrophysiologic study, indicating a role for risk stratification. Appropriate discharges due to ventricular tachyarrhythmias are common in implantable cardioverter defibrillator recipients. Further studies investigating risk factors for adverse clinical outcomes and therapeutic options with larger numbers of patients are needed to adequately address the therapeutic options in patients with congenital left ventricular aneurysms and diverticula.

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